Burdenko's Journal of Neurosurgery
2024, №6, pp. 77–87
https://doi.org/10.17116/neiro20248806177

Giant pericranial sinus with extensive occipital calvarium defect

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ABSTRACT

Background. Giant non-traumatic and non-iatrogenic cranial vault defects are poorly studied due to their rarity. Therefore, diagnosis and analysis of their causes are difficult. In available literature, we found only 4 cases of giant pericranial sinus accompanied by extensive cranial vault defects.

Objective. To present a rare and difficult diagnostic case of cranial vault lesion with non-traumatic and non-iatrogenic extensive bone defect, as well as to propose treatment of this pathology.

The patient presented with complaints of a «growing» parietal-occipital cranial vault defect in November 2022. This defect occurred in childhood. A comprehensive examination including CT angiography of cerebral vessels was followed by surgical treatment (bone resection along skull defect edge with biopsy (aponeurosis, periosteum, bone) and subsequent closure with titanium plate). Postoperative period was uneventful. CT of the skull and brain, including angiography and sinusography, were performed. The follow-up period was more than 10 months with favorable outcome.

Considering anamnesis, clinical and additional diagnostic data, we established giant dominant-type true pericranial sinus.

Keywords: *pericranial sinus, clinical case, differential diagnosis, surgical treatment, rare pathology.*

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TO CITE THIS ARTICLE:
Semenov AV, Semenov AA, Makarova AE. Giant pericranial sinus with extensive occipital calvarium defect. *Burdenko's Journal of Neurosurgery*. 2024;88(6):77–87. (In Russ.). https://doi.org/10.17116/neiro20248806177

Introduction

Pericranial sinus is a rare disease characterized by abnormal communication between dural venous sinuses and extracranial veins through the emissary vein. The last one leads to extracranial venous lake with subsequent osteolysis [1, 2]. Pericranial sinus occurs in people of different ages regardless of gender and previous skull surgery [3].

There are various theories of etiopathogenesis of pericranial sinus. Mastin W.M. divided its causes into congenital, spontaneous and traumatic ones [4, 5]. Muller E. suggested pathogenesis of congenital pericranial sinus and emphasized true angiomas in most cases [6]. Spontaneous sinus is associated with dilation of emissary veins, their pressure on bone tissue and subsequent osteolysis [7]. Traumatic pericranial sinus is associated with damage to emissary vein at the site of exit from the skull [8].

Pericranial sinus usually occurs as a single enlargement of emissary vein in projection of superior sagittal sinus [3, 9]. According to literature data, frontal bone is most often involved in this process, while lesion of parietal and occipital bones is less common [10]. Diameter of vascular anomaly rarely exceeds 1 cm [11]. A typical feature of pericranial sinus is enlargement under Valsalva maneuver [12].

Classification

1. Angioarchitectonics (C. Gandolfo et al.) [13, 14]:
   a) dominant type — brain parenchyma drainage through the veins draining into pericranial sinus;
   b) additional type — partial venous outflow into extradiploic vessels.
2. Reaction of soft tissues within the defect o external compression (J. Volkmann) [13, 15]:
   a) true (enlargement following intracranial pressure increase and disappearance under external pressure);
   b) false (no complete disappearance under compression).

Various studies describe gradual enlargement of pericranial sinus over time [13]. This disease is often asymptomatic and not diagnosed in the early stages [16].

Other causes of spontaneous cranial vault defects include osteogenic lytic sarcoma, intraosseous hemangio-

mas, eosinophilic granulomas, plasmacytoma, dermoid cysts, Ewing sarcoma and Gorham-Stout disease.

A 34-year-old patient came to the Irkutsk City Clinical Hospital No. 3 in October 2022 with complaints of cranial vault defect 7×8 cm in the right parieto-occipital region. There were no clinically significant comorbidities. Skull changes occurred in early childhood and gradually increased over time. This information is evidenced by in-hospital treatment in November 2006 and skull X-ray scan of that time **(Fig. 1)**. A large defect with deformed and partially destroyed outer cortical plate was discovered in parieto-occipital region. There was no surgery due to refusal of the patient and his relatives.

In June 2022, the patient lost consciousness in a sitting position after a bath., Ambulance was called. There was no hospitalization. Neurologist followed-up the patient.

In November 9, 2022, the patient admitted to the Irkutsk City Clinical Hospital No. 3 with complaints of occipital cranial vault defect since birth, weakness, periodic non-systemic dizziness and single loss of consciousness. Local soft tissues were intact, transmitted cerebral pulsation, bulged outward in supine position and sank into cranial cavity in standing position. There were no laboratory abnormalities. MRI (10/27/2022) revealed enlargement of venous confluence and flat vascular formation near bone defect **(Fig. 2)**.

CT (10/28/2022) revealed extensive parieto-occipital defect 7.35×8.72 cm on the right extending beyond the midline to the left **(Fig. 3)**.

To assess superior longitudinal sinus, CT angiography, including imaging of sinuses, was performed on 11/10/2022 (before surgical treatment). We found anomaly of cerebral venous system: dilated veins of Trolard, impaired communication of enlarged venous confluence with superior longitudinal sinus communicating with a wide and flat convexital vascular formation within cranial vault defect filled with blood ("venous lake") **(Fig. 4, 5)**.

To identify possible other foci of osteolysis, we performed X-ray examination of pelvic bones, CT of thoracic spine and chest **(Fig. 6)**. No abnormalities were revealed.

Considering complaints, anamnestic and clinical data, we established the following diagnosis: complex acquired occipital defect of the cranial vault of unspecified genesis, giant pericranial sinus (?), hemangioma (?), eosinophilic granuloma (?), Gorham-Stout syndrome (?).

Considering literature data and risk of traumatic soft tissue damage in projection of vascular anomaly, we established the indications for resection of abnormal bone along skull defect edge with biopsy (aponeurosis, periosteum, bone) and subsequent closure with titanium plate.

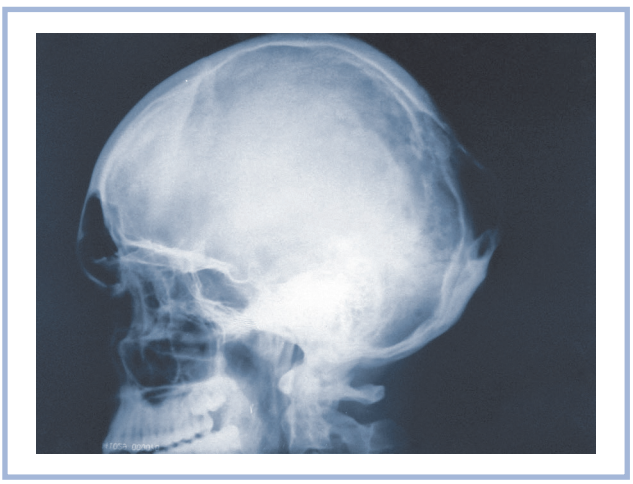


Fig. 1. **Skull X-ray scan, lateral projection (11/16/2006, 18 years old).**
Parieto-occipital bone defect with partially destroyed outer cortical plate.

Surgery (11/22/2022)

We incised and separated skin and aponeurosis from periosteum. There was blood accumulation under perios-

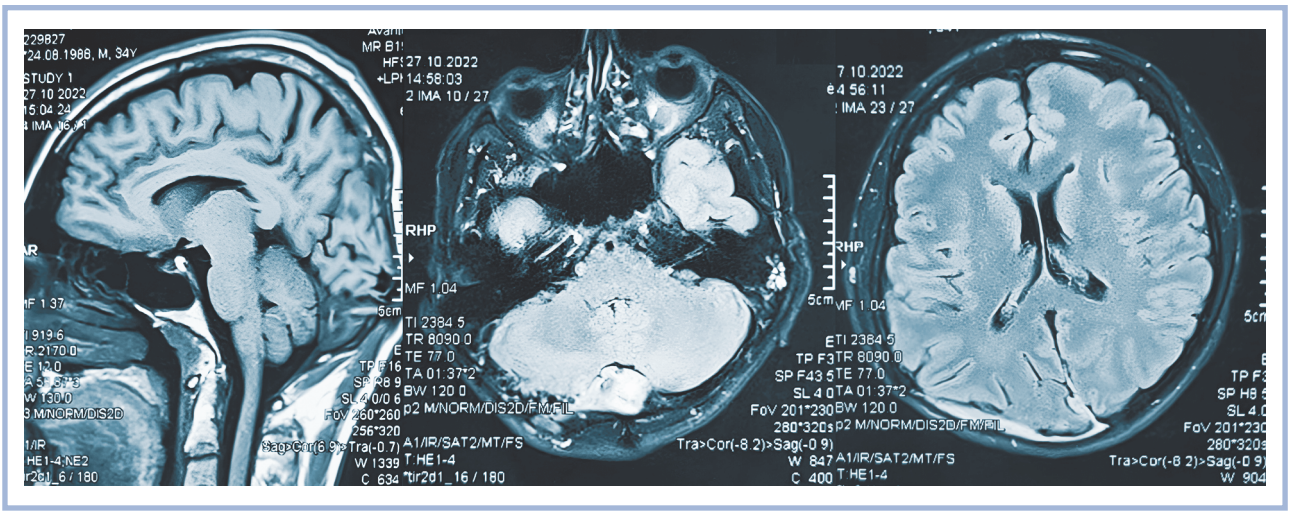


Fig. 2. **Sagittal and axial MR scans (10/27/2022).**
Enlargement of venous confluence and flat vascular formation near bone defect.

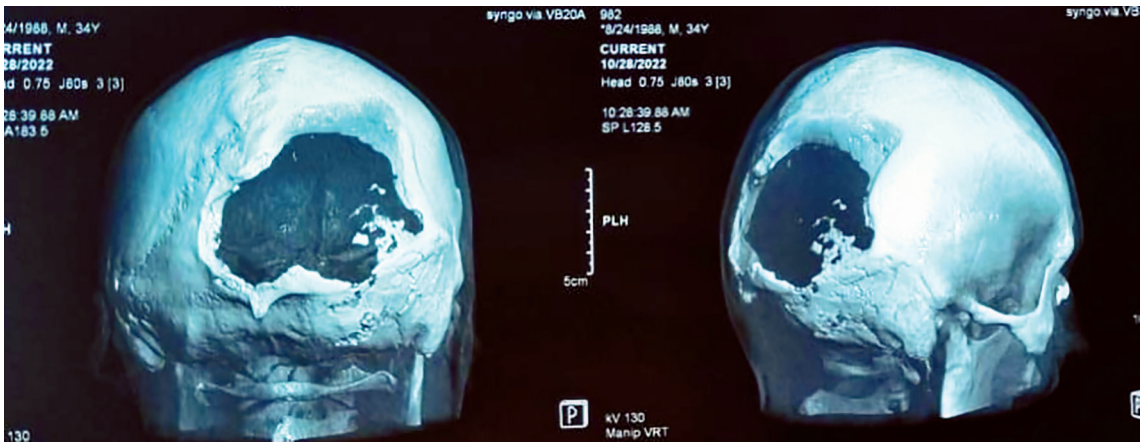


Fig. 3. **CT with 3D bone reconstruction (10/28/2022).**
Extensive right-sided parieto-occipital bone defect extending to the left beyond the midline.

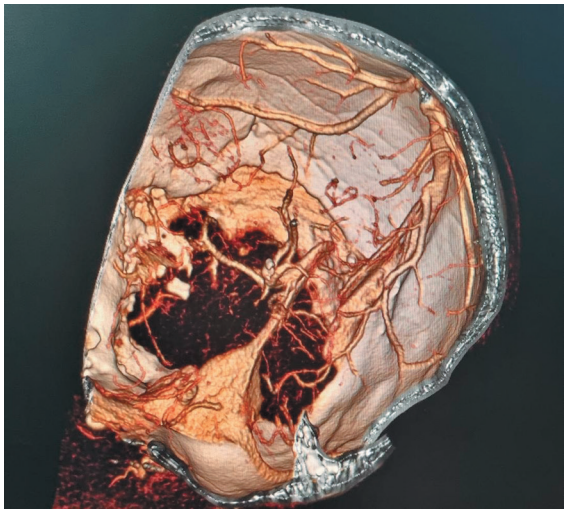


Fig. 4. **CT angiography of the brain, sinusography phase, 3D reconstruction (11/10/2022).**
Sinus rectus is enlarged at the level of venous confluence. Communication of superior sagittal sinus with venous confluence is impaired at the level of posterior third. Veins of Trolard are enlarged. There is phenomenon of venous lake in the area of bone defect.

teum over the entire area of bone defect (7.35×8.72 cm) in the form of extensive "venous lake". This finding corresponded to preoperative sinusography data **(Fig. 4, 7)**. We skeletonized the defect edges along the perimeter 1 cm away from the edge. Mobilization of bone defect edge towards dura mater was accompanied by venous bleeding that prevented "refreshing" of edges. To separate abnormal bone tissue of defect edge from intact skull, we performed channel-like bone resection along the defect edge (5 mm wide). Finally, we visualized diploic veins with profuse bleeding and performed hemostasis using wax. As resection progressed, "venous lake" bulging into surgical wound increased. This indicated participation of diploic veins along the perimeter of the defect in blood drainage from the "lake" **(Fig. 7)**. Preoperative sinusography did not establish this fact.

Compression of bulging wall of "venous lake" reduced its volume to baseline status, but it increased again later. In this way, we could assess integrity of other venous anastomoses under destruction of diploic veins. Parietal canal resection was bilateral through the midline to superior nuchal line. We left 3 bone bridges in parietal regions fixing the separated bone along the defect edge. There was no resection in occipital region due to the risk of disruption of venous outflow from venous "lake". Next, we fixed an appropriate titanium plate to the skull using cortical screws. Wound closure in layer-by-layer fashion was accompanied by epidural drainage. Blood loss was up to 400 ml, blood transfusion was not performed.

Biopsy of bone tissue of defect edge, periosteum and aponeurosis in projection of venous anomaly was performed. Biopsy of "venous lake" walls was not performed due to the risk of life-threatening bleeding. Analysis presented ambiguous result **(Fig. 8)**. Bone tissue of parietal bone defect edge had signs of decalcification. Periosteum and aponeurosis were represented by dense fibrous connective tissue with bundles of skeletal muscles. Specimen of occipital bone defect edge demonstrated small-caliber venous vessel growths with signs of perivascular inflammation. These findings excluded malignant nature of disease. However, they could be observed in pericranial sinus and Gorham-Stout disease [17, 18].

Postoperative period

Postoperative period was uneventful. The patient was discharged after 14 postoperative days. An-



Fig. 5. **CT angiography of the brain, sinusography phase, axial, frontal and sagittal scans (11/10/2022). Axial scans at the levels of "venous lake" (a) and enlarged venous confluence (b).**

ticoagulant therapy was administered for 2 months after discharge (enoxaparin sodium 40 mg 2 times a day for 1 month subcutaneously with control of clotting parameters once a week). The follow-up examinations revealed clinical improvement with relief of preoperative symptoms.

The patient underwent postoperative imaging for control of circulation in vascular anomaly **(Fig. 9—11)**. We observed gradual normalization of circulation with partial thrombosis of dilated veins, disappearance of venous confluence enlargement and reduced collateral blood flow through the veins of Trolard.

Discussion

We retrospectively analyzed diagnostic data to verify the diagnosis.

Osteogenic lytic sarcoma can lead to skull defect with clear soft tissue thickening, and this volume does not decrease in horizontal position [19]. Magnetic resonance imaging, contrast-enhanced CT and histological examination of tumor are used for differential diagnosis. Osteosarcomas accumulate contrast agent unevenly. Histologically, these tumors are represented by spindle-shaped cells with pathological mitoses and nuclear hyperchromia,

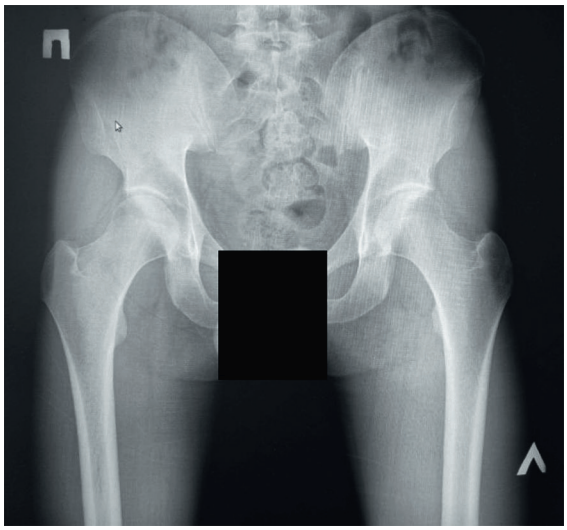


Fig. 6. **Pelvis X-ray (11/14/2022). No bone destruction is detected.**

as well as needle-shaped fan-shaped shadows in periosteal tissues [20, 21].

Intraosseous hemangiomas can also cause osteolysis. They are characterized by debut in childhood, lesion of several anatomical areas of the head and infiltrative growth [22].

Intraosseous eosinophilic granuloma is bone tumor characterized by infiltration and proliferation of pathological Langerhans cells with potential osteolysis [23]. There are high density and small sizes of bone defects. This diagnosis can be unambiguously confirmed by histological examination of tumor. We can observe Langerhans cells with polymorphic nuclei, clusters of eosinophils and histiocytes [24].

Cranial vault plasmacytoma is most often a sign of multiple myeloma. Its histological signs are atypical plasma cells with pathological mitoses and nuclear hyperchromia [25].

Dermoid cyst is a benign tumor with fibrous capsule consisting of ectoderm and mesoderm precursor cells. Dermoid cyst is capable of causing osteolysis. MRI visualized dermoid cyst as a heterogeneous tumor replacing bone. Dermoid cyst accumulates contrast agent well. It contains oily fluid with fragments of ectodermal and mesodermal tissues [26]. Histological picture is represented by keratinized multilayered squamous epithelium containing hair follicles, sebaceous and sweat glands [27].

Ewing tumor leads to multiple small foci of destruction of diploic and compact substance. This tumor usually occurs in young boys [21].

In our case, histological analysis of occipital and parietal bones, as well as aponeurosis and anamnestic data excluded the majority of suspected diagnoses (osteogenic lytic sarcoma, hemangioma, eosinophilic granuloma, plasmacytoma, dermoid cyst, Ewing tumor). However, baseline data did not allow Gorham-Stout disease to be definitely excluded [17].

Gorham-Stout disease is a rare disease of unknown etiology characterized by osteolysis and proliferation of vas-

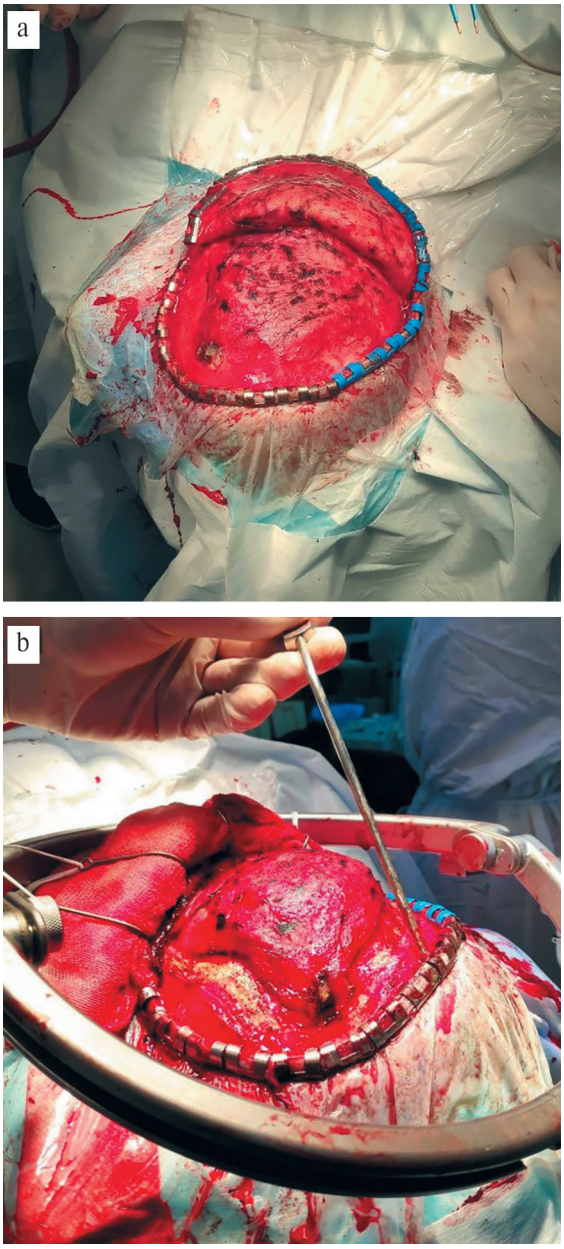


Fig. 7. **Change in the overall appearance of the "venous lake" due to diploietic vein destruction.**
a — "venous lake" before destruction of diploic veins (intraoperative image); b — "venous lake" after partial destruction of diploic veins; increased volume is visualized (intraoperative image).

cular structures [28]. This disease can affect one or more bones including skull, spine, upper and lower limbs, pelvis. Gorham first described this disease in 1954, and Stout reported this disease a year later.

Differential diagnoses also included pericranial sinus. However, this disease very rarely proceeds in this fashion. In available literature, we found only 4 case reports on giant pericranial sinus accompanied by extensive cranial vault defects.

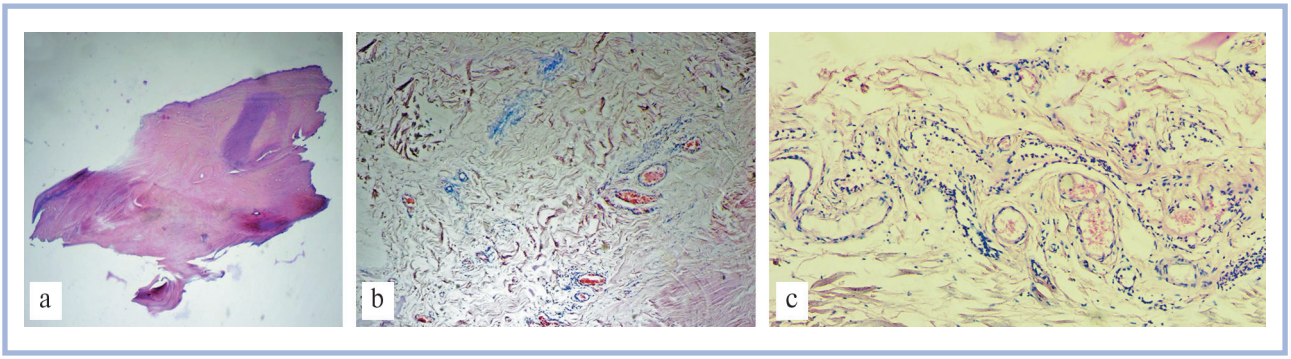


Fig. 8. **Histologic picture of the operative material taken from areas adjacent to the anomaly.**
a — bone tissue of defect edge (parietal bone), decalcification, hematoxylin and eosin staining, ×40, bone tissue is represented by compact bone substance, bone plates are tightly adjacent to each other, osteons and lacunae with osteocytes along the periphery are visualized; b — aponeurosis, hematoxylin and eosin staining, ×40, fragments of fibrous connective tissue with areas of hyalinization and bundles of skeletal muscles are visualized; c — bone tissue of defect edge (occipital bone), hematoxylin and eosin staining, ×100, proliferation of small-caliber venous vessels like vascular venous malformation filled with blood, perivascular inflammation and fragments of the periosteum are visualized.

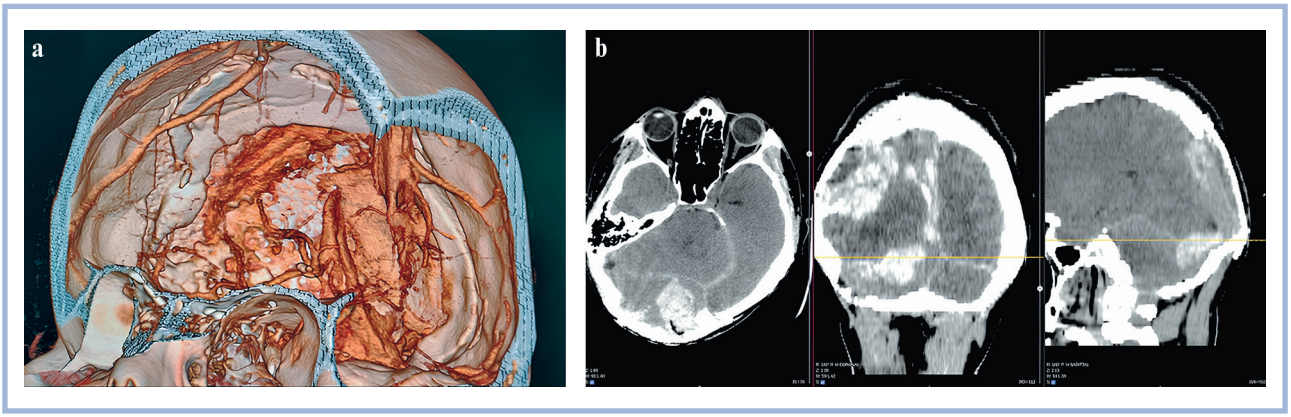


Fig. 9. **Sinusogram and MSCT of the brain.**
a — sinusography, 8 days after surgery (11/30/2022); b — CT without contrast-enhancement, partial thrombosis of venous lake (11/30/2022).

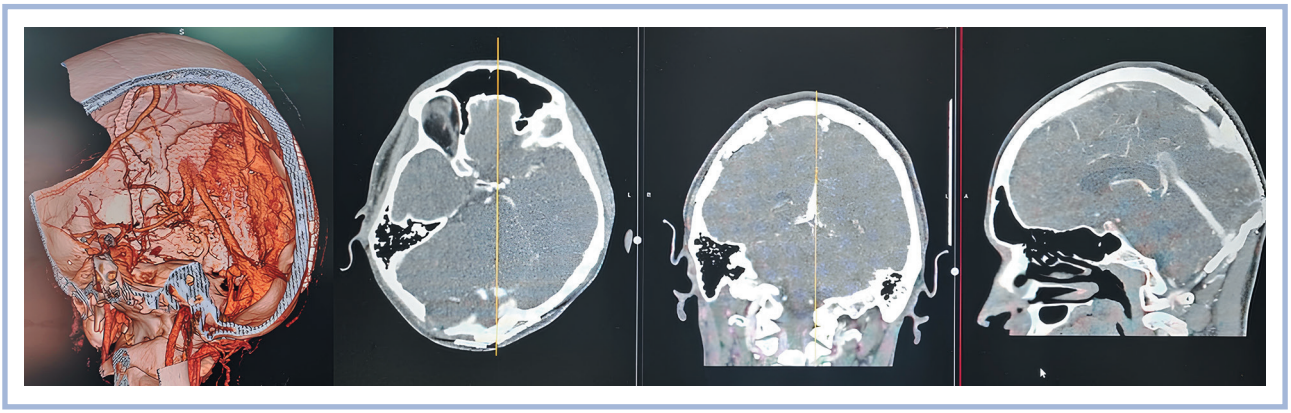


Fig. 10. **Sinusography, 3 months after surgery (02/01/2023); shrinkage of venous confluence, partial recovery of blood flow between superior longitudinal sinus and venous confluence.**

Thus, we differentiated Gorham-Stout disease and pericranial sinus in our case **(Table)**.

Thus, it is difficult to differentiate these diseases despite the obvious difference in biological nature. The diagnosed "venous lake" within the bone defect combined with anomaly of venous sinuses, as well as no osteolytic lesions of other bones indicated a giant pericranial sinus. Constant changes in intracranial pressure, as well as higher pressure inside "venous lake" and diploic veins when changing body position or under local pressure on occipital tissues in supine

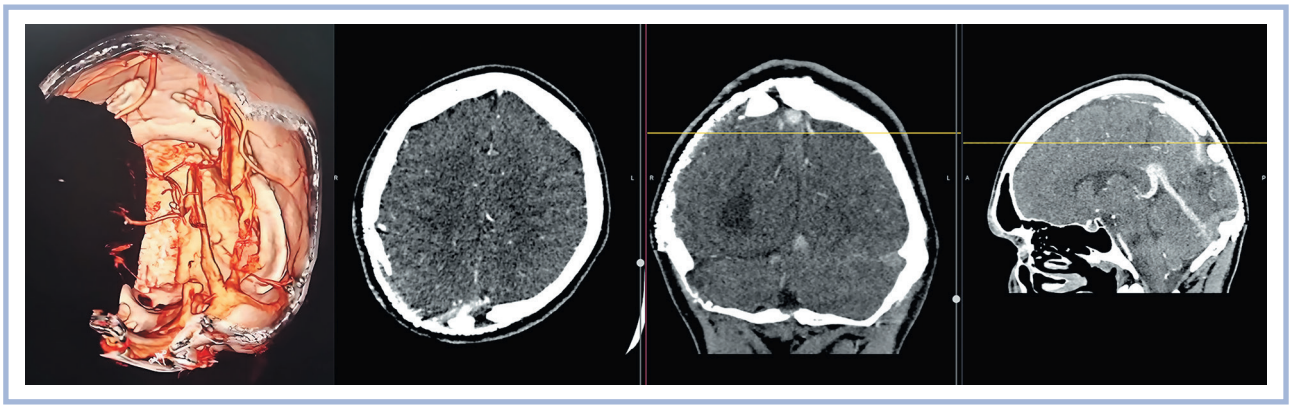


Fig. 11. **Sinusography, 7 months after surgery (05/30/2023); shrinkage of venous confluence, reduced volume of collateral vessels.**

Differential diagnosis of Gorham-Stout disease and pericranial sinus

Signs in our patient	Signs of Gorham-Stout disease	Signs of pericranial sinus
Large bone defect	+	+/–
Venous lakes	–	+
Asymptomatic course	+	+
Enhancement of diploic vascularization	+	+
Long-term progression	+	+
Pathological proliferation of vessels within the bone with bone tissue replacement	+	+
Bone defect enlargement over time	+	+

position could contribute to progression of skull osteolysis around the "lake". A "vicious circle" was possible (larger "venous lake" contributes to slower venous drainage with subsequent pressure increase inside the lake). Considering literature and clinical data, we established true (according to J. Volkmann) dominant-type (according to C. Gandolfo) giant pericranial sinus with gradual enlargement.

This vicious circle may be based on direct relationship between higher pressure inside "venous lake" and its volume. The last one may be due to individual anatomical features of venous sinuses and changes in blood flow through these sinuses due to their thrombosis, damage or structural anomalies. Perhaps, surgery of venous sinuses leading to normalization of venous circulation and lower pressure in "venous lake" should underlie treatment of this disease. According to the literature, dominant-type pericranial sinus is a contraindication for detachment from drainage vessels due to possible complications such as venous hypertension and venous cerebral infarctions [13, 14, 29]. Nevertheless, surgical intervention was necessary due to high risk of damage to venous lake in everyday life with extensive bleeding and death. In addition, malignant nature of disease was not completely excluded. In this case, detachment of damaged bone from intact tissues could stop osteolysis. Surgical strategy is not ideal in this case. However, it contributed to clinical improvement, positive correction of hemodynamics in venous sinuses and shrinkage of "venous lake". Perhaps, this will lead to disappearance of "vicious circle" and recovery in the future. We hope that further follow-up will confirm our hypothesis.

Conclusion

Pericranial sinus is a rare disease whose diagnosis and treatment require analysis of literature data. When making this diagnosis, it is important to differentiate pericranial sinus with other multiple skull lesions including Gorham-Stout disease.

At the same time, to date, there is no standard management of patients with this diagnosis. In this report, we present a possible variant of pathogenetic surgical treatment of pericranial sinus, i.e. disconnection with some diploic veins leading to partial thrombosis of vascular anomaly, decrease of internal volume and pressure with normalization of blood flow.

Author contribution:
Collection and analysis of data — Semenov A.A.
Examination and treatment — Semenov A.V.
Surgery — Semenov A.V.
Histological analysis — Makarova A.E.
Writing the text — Semenov A.A.
Editing — Semenov A.V.

The authors declare no conflicts of interest.

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Received 17.09.2023
Accepted 20.09.2024

COMMENT

The manuscript is devoted to diagnosis and surgical treatment of extremely rare disease, i.e. acquired spontaneous progressive extensive cranial vault defect. The authors analyze the literature data devoted to this issue, as well as describe various diseases requiring differential diagnosis in these patients. Neuroimaging assumed secondary nature of progressive cranial vault defect following vascular anomaly. Histological examination and analysis of skull bones were performed to clarify the diagnosis and exclude primary bone lesions. Considering diagnostic data, we diagnosed a giant pericranial sinus. A theory of development and progression of bone defect associated with persistent blood "lake" is described. Surgical treatment was preventive regarding traumatic damage to this vascular anomaly. Postoperative follow-up revealed clinical improvement after skull repair.

Thus, this report demonstrates an extremely rare disease, i.e. giant pericranial sinus with secondary progressive cranial vault defect, as well as one of the possible surgical options for this disease.

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